

SPECIAL ISSUE

Facilitating Motor Learning in a Child with a Chromosome Translocation: Applications of Quantitative Surface Electromyography

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Physical rehabilitation typically conjures up images of an adult in physical therapy to recover from an accident or injury. Children are also the unfortunate victims of strokes, head injuries, and developmental disorders. In this case study, a child with a rare genetic disorder is helped to learn to stand using a modification of surface electromyography, quantitative surface electromyography (QSEMG). Over a period of twelve 1-hour sessions, she learned to stand with minimal assistance. Achieving these types of goals is of critical importance for the development of maximum independence in performing activities of daily living.

Introduction

In past articles (Bolek, 1998, 2001, 2006, 2010, 2012, 2013), the first author explored the use of surface electromyography (SEMG) in the rehabilitation of motor function subsequent to a brain injury. This technique evolved over the years into what became known as quantitative surface electromyography (QSEMG), whereby the SEMG signal is partitioned into its component parts based on a predetermined algorithm. It has been observed in the popular press that only a moment in time separates each one of us from a catastrophic injury. No one expects to be the victim of a drunk driver, a mechanical malfunction during a plane takeoff, the sudden loss of function while at work due to a stroke, or the birth of a child with significant disabilities. In a very real sense, psychological health is partly dependent on a well-functioning defense system that keeps at bay the many realities of daily life. Unfortunately, resources to treat these medical conditions are often the first cut during times of fiscal budget cutting. Consequently, methods to help restore function to those afflicted must demonstrate cost-effectiveness in order to be covered by insurance. QSEMG is one method of attaining these goals. For most, “physical rehabilitation” conjures up images of an adult

hard at work in a physical therapy program, for example, relearning to stand after suffering a stroke. The challenge is increased enormously when we deal with those patients who, for a variety of reasons, cannot or will not be cooperative patients. In this case study, we tracked the progress of “Ann,” a now 4-year-old girl, through a QSEMG-based treatment program.

Medical History

Ann was the product of a full-term pregnancy that was complicated by group B streptococcus infection and preeclampsia, which occurred at the time of delivery and was treated with intrapartum antibiotics. There were no other prenatal risk factors. Shortly after birth, she began having problems with feeding and developed frequent episodes of projectile emesis, reflux, and poor weight gain. At about 6 months, she showed signs of hypotonia based on poor head control and progressive lethargy. At 7 months, she developed a seizure that required hospitalization and an extensive neurologic workup. During that hospital admission, she underwent extensive metabolic evaluations, which showed no specific diagnoses; however, a magnetic resonance imaging study revealed some degree of brain asymmetry. A video electroencephalogram suggested multifocal areas of abnormalities. She continued to have multiple episodes of vomiting thought to be partially due to gastroesophageal reflux. This diagnosis was confirmed by a pH probe study. There was no history of recent fevers, chills, anorexia, or unexplained weight loss. Socially, Ann was minimally interactive with either those in the room or the environment in general. She showed a high degree of tactile sensitivity and anxiety. Her specific diagnosis was listed as “deferred,” but general descriptors included failure to thrive, gastroesophageal reflux, developmental delay, hypotonia, musculoskeletal weakness, seizures, visual difficulties, noncommunicative, microcephaly, and mild

craniofacial dysmorphisms. Subsequent genetic testing revealed an X;2 balanced translocation, which is a rearrangement of material between 2 different chromosomes. However, the assessment was unable to identify an abnormality associated with Ann's translocation that could be the underlying cause of her medical problems. In effect, the medical community has been unable to establish the origin of her developmental problems, and her final diagnosis remains a mystery.

Academically, she is in pre-kindergarten. She enjoys horseback riding and swimming and does participate in physical, occupational, speech, and language therapy. She is not very interactive with those around her. She is not autistic but prefers to be minimally interactive. She displays good eye contact. Touch not initiated by her is not well received. She enjoys playing cause/effect as well as simple musical games on her iPad. She displayed about the same level of interaction with those in the room, be it her mother, a nurse, therapist, or doctor. She prefers self-directed activities; thus, it is difficult to engage her in a therapist-directed/-selected activity. Ann has had many therapeutic interventions the past 4 years of her life, including physical ("land" therapy and aquatic environment), occupational (fine motor and feeding skills), speech therapy (including augmentative communication), and hippotherapy (rehabilitative and therapeutic activity aided by a horse). She has been using a "sit to stand" stander for the past 3 years, and she is able to locomote independently with a KidWalk gait trainer. She presents with a flexor-withdrawal reflex upon standing or weight-bearing activities. She also presents with a severe form of hypotonia throughout her trunk and all extremities. Compressive garments, thought to provide the trunk with maximal proprioception in all directions, had been tried out on several occasions to assist with her upper- and lower-extremity control. Because of the lack of expressive communication, it has never been known how much this child is able comprehend or is able to be motivated.

Development of a Treatment Plan

Assisting a patient in the relearning of motor skills requires some basic skill sets to be intact. First, there must be the ability to acknowledge cause and effect. For a very young child, this may be demonstrated by the active pressing of a switch to activate a toy. Second, there must be the ability, once the cognitive schema is formed, to use motor skills to act on the environment. Those unfortunate enough to have suffered an injury that results in a "locked-in" syndrome are aware of their environment but are minimally able to respond. Third, there has to be some minimal motivation to

perform the movement. Fourth, at some point, some patients will need to discover the use of proprioception. Proprioceptors are neuromuscular joint receptors that register stimuli such as stretch, tonicity, and movement within muscles. An example of proprioceptive awareness is knowing where one's body is in space. Patients lacking this awareness may have difficulty remaining sitting upright on a bench simply because they do not have a sense of where their pelvis is in relation to the bench. Inactive or minimally active proprioceptors are commonly found in children with hypertonia, as in Ann's case.

Implementation of the Treatment Plan

Ann was lacking several skill sets. There were opportunities to work on sitting, standing, reaching, use of a motorized wheelchair, and the sit-to-stand function. Standing was selected as the target because a common concern as a child ages is the natural increase in weight, although standing was her least preferred activity. If a child can learn some independence in standing, activities of daily living (such as toileting, grooming, brushing teeth) will result in less wear and tear on the back of the primary caregiver. Of the four requirements mentioned earlier to participate in this kind of treatment, all four were met, with the exception of motivation. With Ann's limited social interaction, it was not readily apparent whether she would find the activity of standing in and of itself rewarding.

The details of the treatment methodology have been described previously (Bolek, 2001, 2010, 2012). In QSEMG, the focus is to internalize the correct muscle pattern recruitment within the constellation of muscles rather than relying on only one or two muscles. Each target muscle is selected by the clinician for its relevance to the functional goal. When this constellation of muscle groups is on target (i.e., the thresholds met), positive feedback in the form of a video reward is activated. Typically, some muscles need to be activated and some relaxed. For example, upright sitting may be enhanced by targeting the lower trapezius to be active with minimal use of the upper trapezius. Failure to maintain any muscle at the therapeutic threshold terminates the reward. In Ann's case, the muscles targeted were the bilateral gluteus maximus and quadriceps, the primary muscles needed to maintain a standing posture (Fischer & Houtz, 1968; Karlsson & Jonsson, 1965). The gluteus maximus serves as an anchor for the pelvis during standing, whereas the quadriceps enable the patient to fully extend the legs. A range was set up for all four sites such that overuse/underuse was the "no-go" criteria. In other words, activation of all four muscles, to a point, activated the video reward. Setting a range for the patient-generated effort as

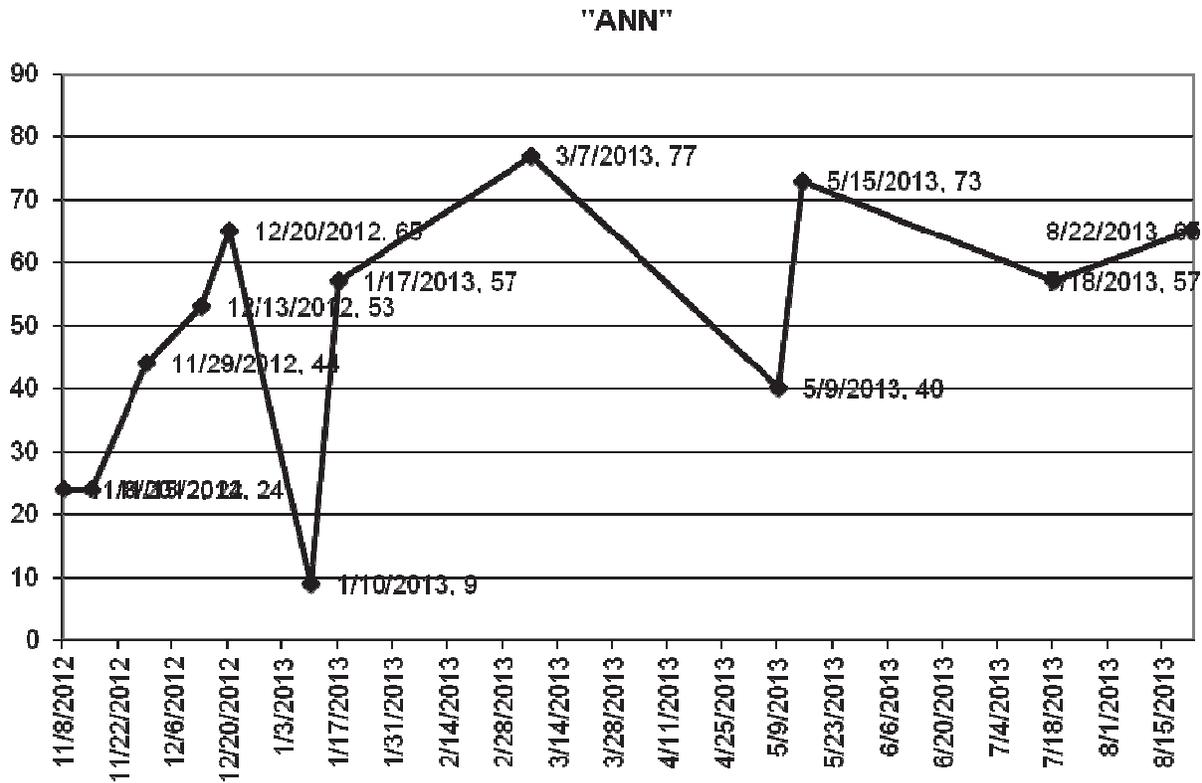


Figure 1. Ann's progress over 12 sessions. The first two values overlap. Both 11/8 and 11/15 are 24%.

measured by the SEMG signal is the preferred method for a child this age because of the tendency, when tired or frustrated, to squirm or tense the whole body. Disposable pregelled electrodes (GS-27, Bio-Medical Instruments, Inc., Warren, MI) were applied after the skin was abraded with alcohol. The bandwidth of the encoder was set at 20 to 500 Hz.

There were several challenges to be addressed in the development of a treatment program. As mentioned earlier, Ann was tactilely defensive; therefore, the typical hands-on support given when working on standing was not well received. She was not one who was easily engaged socially. She had perseverative tendencies manifested by some difficulty in transitioning between tasks. It was not clear how she would respond to the sudden turning on/off of the video based on the motor pattern she was displaying. There was no way to ask her opinion on the kind of reward she would enjoy (as far as the choice of the movie) and receive a communicative response. After considering these possible impediments to progress, it was decided to proceed with the program. Up to this point in her therapy, the functional gains achieved had been slow in coming. The first three sessions were carefully evaluated for signs of progress and indicators that the program was too demanding for her age and disability level. It was clear after the first session that

she recognized the relationship between “tall standing” and onset of the video. She was not happy when the video ended due to not meeting the thresholds set for muscle use but could maintain an even level of participation throughout the 1-hour sessions.

She continued to have largely undefined gastrointestinal issues throughout the treatment that caused generalized discomfort. For this and other reasons, her emotional state during any given session varied greatly. Coexisting alongside these issues was the natural tendency for a child this age to test the limits by simply refusing to cooperate. Various behavior management techniques were used to address these episodes when they occurred (turning away from her with no eye contact and placing her in a supine position on the floor until the tantrum stopped).

Outcome

Figure 1 displays Ann's progress over 12 nonconsecutive sessions spanning 10 months. Interruptions were primarily due to patient illness. Ann had a history of periodically occurring seizures. She also would have periods of undiagnosed abdominal discomfort. Nevertheless, if one removes the two low scores from the graph (1/10/13, 5/9/13), there is a consistent pattern of upward progression from a low of 24% on 11/8/12 to 65% on 8/22/13.

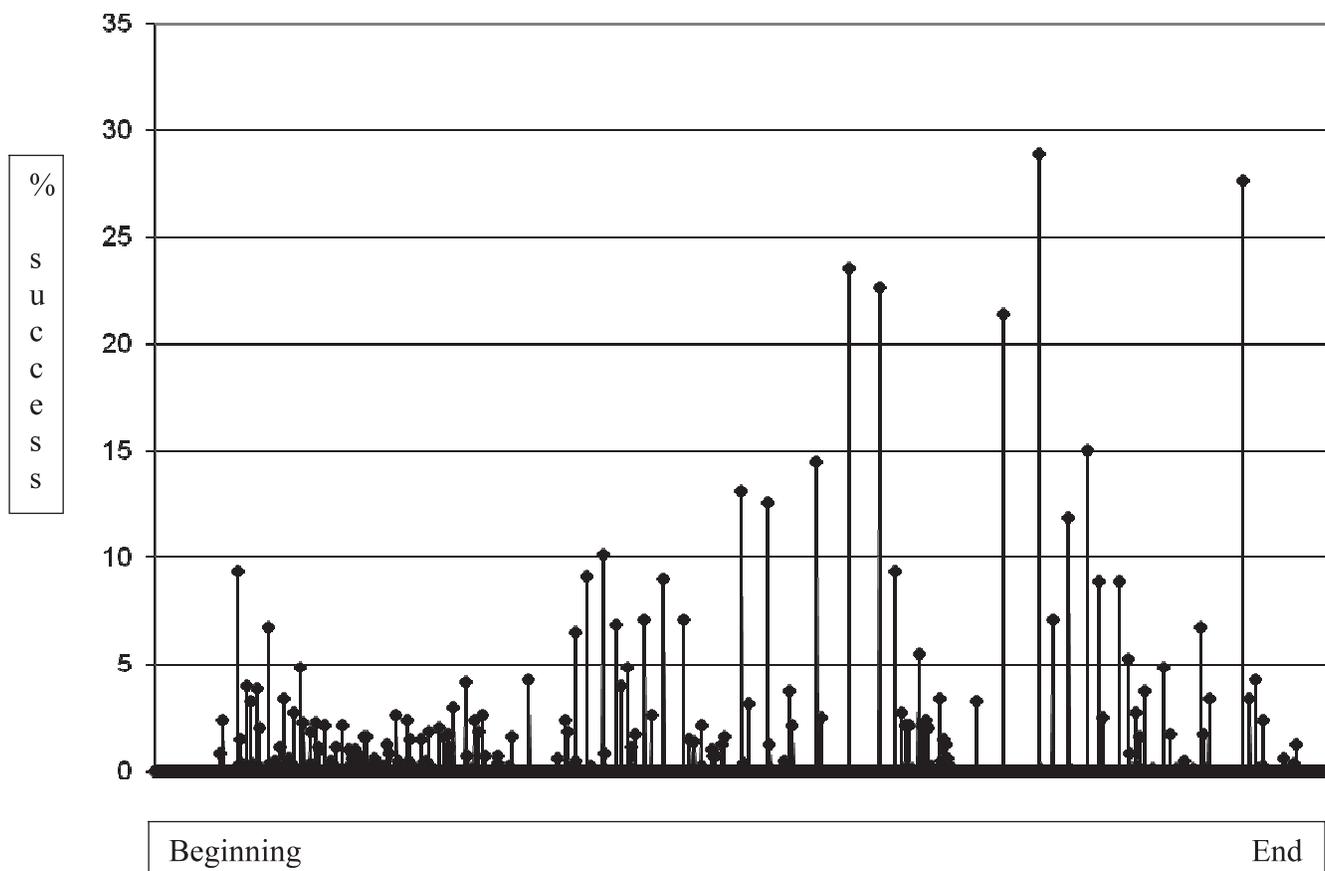


Figure 2. December 13, 2012. Session progress, 50 minutes.

Moreover, this increase was accompanied by parental reports of greater participation on the part of Ann during toileting and dressing. Her ability to stand increased in both quality and duration. A side benefit of the program was increased tolerance for a delayed reward. A critical aspect of the program was linking a verbal cue with the learned performance. The moment she mastered proper standing (all thresholds met), she was told, “good standing.” By association, this verbal prompt, after repeated practice, could be used as a cue to stand correctly outside of the session, thereby continuing the treatment program beyond the confines of the motor control laboratory.

Prior to starting the QSEMG program, Ann was unable to even minimally weight bear through her lower extremities in standing, without full coverage bracing or donning knee immobilizers. She was also unable to functionally engage in play with toys using her hands. She was able to do so after 4 weeks of treatment. The establishment of a base of support (gluteal muscles) typically allows the patient to explore upper-extremity functioning. All attempts to enhance her ability to stand

and bear weight volitionally had not been successful until she started the QSEMG program.

Discussion and Summary

In an ideal situation, a patient that has sustained loss of motor function would be motivated to change, if he or she is old enough to realize the implications of continuing to use motor patterns that stress ligaments and joints, has the endurance to cope with the demands of therapeutic sessions, and has the financial capacity to continue the treatment long enough for a successful outcome. Unfortunately, in our society, benefits for the disabled are often the first to be cut fiscally, because the lobbying efforts are miniscule compared with other programs. SEMG, specifically QSEMG, can provide a treatment method that is cost-effective and able to generate statistical outcomes both within and across treatment sessions, with permanent improvement in functional outcomes. The treatment is especially applicable for those patients who are very young, difficult to motivate, and not of the age at which they can appreciate the consequences of continuing to use maladapt-

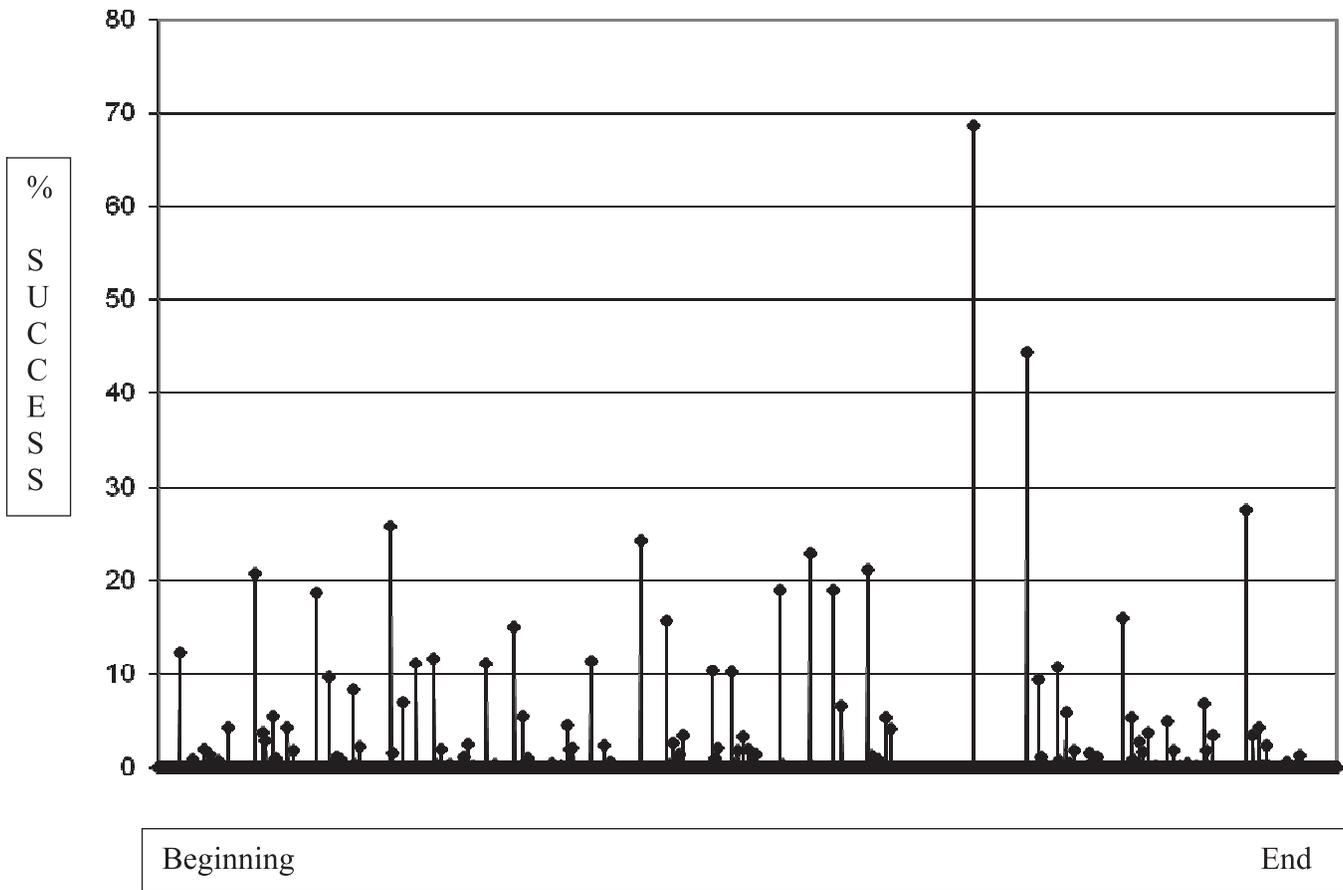


Figure 3. August 22, 2013, session progress, 50 minutes.

tive motor patterns. For this patient, the treatment term spanned 9 months. One advantage of QSEMG is the ability to track progress within and across sessions. Although Figure 1 shows the progress across sessions, Figures 2 and 3 show the progress within a 50-minute session. Of interest is that the December 13th graph (2012) shows a maximum consecutive standing time of about 28 seconds. On August 22, 2013, this has increased to nearly 70 seconds. This type of data helps justify continued treatment.

There were several characteristics of Ann’s case that mitigated against treatment success, such as her young age, tactile defensiveness, limited social interaction, ongoing profound medical issues, and absence of a clear medical diagnosis. QSEMG could be used in similar cases to improve the quality of life of those in need of rehabilitation.

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